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MRA Detection of Vascular Occlusion in a Child with Progeria

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Summary: We report a case of progeria and the utility of visualizing the cerebrovascular anatomy by using MR angiography. A 4-year-old child with Hutchinson-Guilford syndrome developed symptoms of ischemia and MR angiography showed bilateral occlusion of internal carotid and vertebral artery origins; the anterior spinal artery was prominent.

Index terms: Progeria; Brain, magnetic resonance; Pediatric neuroradiology; Magnetic resonance angiography (MRA)

Hutchinson-Gilford syndrome (progeria) is associated with several features of premature aging. It is manifested by growth retardation, characteristic facies, loss of hair and subcutaneous fat, restricted joint mobility, and severe atherosclerosis (1). Complications of atherosclerosis include cerebrovascular disease with resultant transient and permanent neurologic sequelae. The significant morbidity and mortality associated with intraarterial angiography in this population lessens its utility in the radiologic assessment of cerebrovascular complications. We report the application of a noninvasive method to visualize the larger cerebral vasculature, in a 4½-year-old child with progeria and recurrent neurologic dysfunction.

Case Report

At age 4 years, this boy with Hutchinson-Gilford syndrome (progeria) developed transient episodes of headache, drooling, and right arm weakness. Within 1 month, he had a right-sided tonic seizure followed by right arm and leg weakness. Examination revealed bilateral carotid and left ocular bruits, decreased left carotid artery pulse, grade I-II/VI systolic murmur, mild lethargy, no facial asymmetry, flaccid and weak right leg and right arm, bilaterally brisk deep tendon reflexes, and a right extensor plantar response. A CT scan of the head taken at his hospital admission showed right parietooccipital and left frontal subdural collections and an acute left posterior parietal infarction. Seizures that recurred were controlled with phenytoin and carbamazepine therapy. On day 3 after admission, parenchymal magnetic resonance (MR) imaging showed bifrontal and right posterior parietal subdural fluid collections, diffuse periventricular white matter and basal ganglia ischemic disease, and a right posterior parietal infarct. (Fig. 1). MR angiography (MRA) demonstrated bilateral occlusion of the proximal internal carotid arteries and origins of the vertebral arteries, with multiple cervical collateral vessels reconstituting the cavernous left internal carotid and both cervical-vertebral arteries. The anterior spinal artery was prominent and was therefore also assumed to be a source of collateral flow (Fig. 2). There was generalized decreased intracranial flow for a child of this age and markedly diminished flow in the right anterior cerebral artery and branches of the middle cerebral arteries. Increased flow signal was present in both ophthalmic arteries, again thought to be acting as collaterals (Fig. 3). Coumadin treatment was started for presumed embolic disease. His right hemiparesis slowly improved but transient episodes of left-sided weakness occurred. MR studies 6 months after his hospital discharge showed bilateral frontal and parietal subdural collections, increased prominence of sulci and ventricles, periventricular white matter signal abnormalities, and biparietal and right frontal cortical infarctions. The MRA study was unchanged.

Discussion

Premature atherosclerotic disease of the cardiovascular system is well documented in progeria patients and is a major contributing factor to their shortened life spans. In a review of 60 cases, acute myocardial infarct was the most common cause of death, followed by chronic myocardial infarction (1). Few other causes of death have been reported. The cause of the premature atherosclerosis in progeria remains unknown. Pathologic studies have found premature subintimal fibrosis (2). One theory surmises that the endothelium is incapable of adequately restoring itself with resultant atherosclerotic plaque

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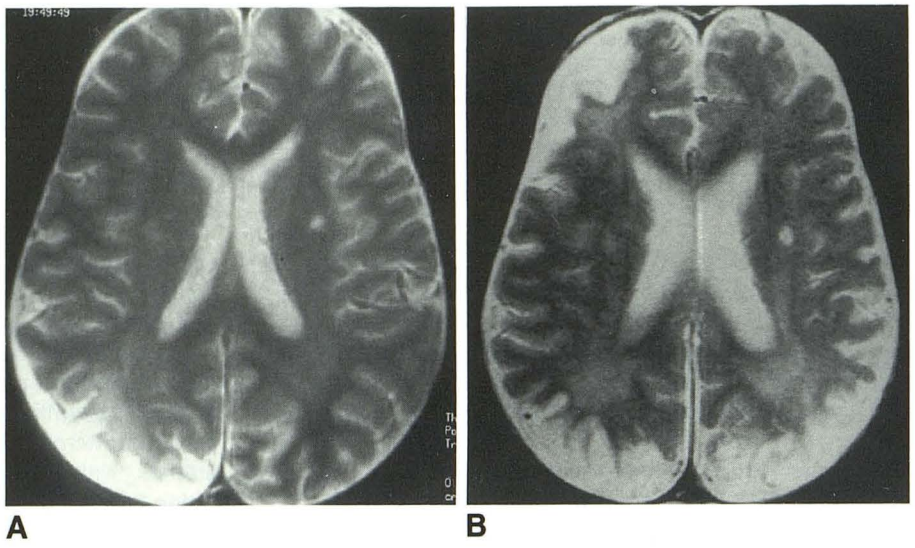


Fig. 1. MR of the brain.

A, Bilateral extraaxial fluid collections, a left parietal infarct, and mild white matter changes are present.

B, Interval study 6 months later shows progression to biparietal and bifrontal cortical infarcts, bilateral deep white matter ischemic changes, and mild ventriculomegaly (2000/90, TR/TE).

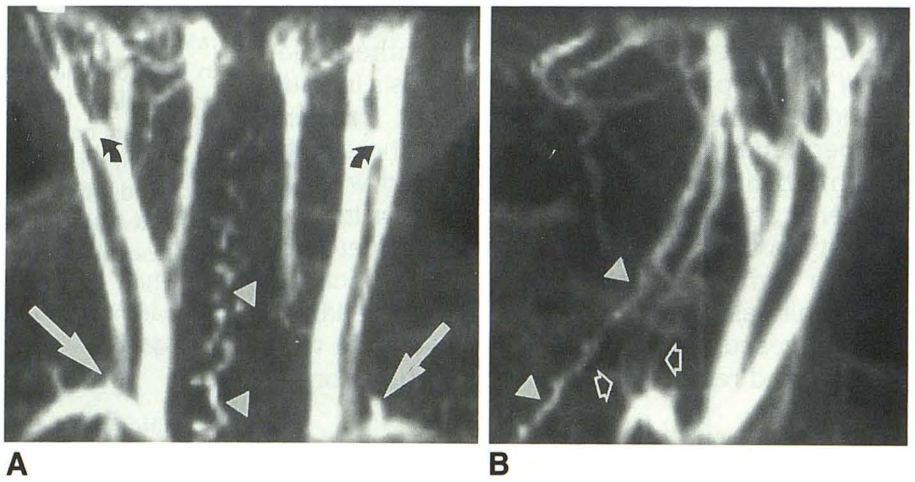


Fig. 2. Three-dimensional time-of-flight MRA of the neck.

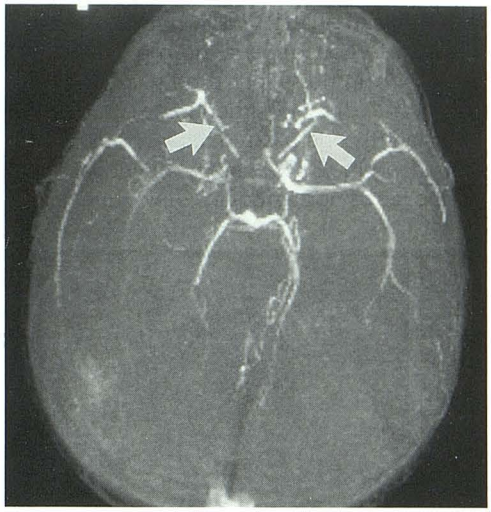
A, The anteroposterior view shows apparent occlusion of the internal carotid artery origins (*curved arrows*), apparent occlusion of vertebral artery origins (*straight arrows*), and tortuous anterior spinal artery (*arrowheads*).

B, Right-anterior-oblique view suggest reconstution of the distal cervical vertebral artery by branches of the thyrocervical trunk (*open arrows*). The anterior spinal artery, which is assumed to give collateral flow, is seen posteriorly (*arrowheads*). (3-D fast imaging with steady precession (TR/TE/flip angle) 40/13/15°, no venous presaturation).

formation. Support of this theory comes from studies of cell cultures that show decreased reparative ability for DNA breaks caused by x-rays (3).

Cerebrovascular disease has not been well described in progeria. To our knowledge, this is one of only three documented cases in children with progeria. The other cases include a 7-year-old boy with a large left hemisphere infarct (4) with complete occlusion of the left internal carotid artery by angiography; and a 9-year-old girl with prominent cardiovascular problems and angiographic findings of severe atherosclerotic disease of the vertebral arteries and complete left internal carotid artery occlusion (5).

Our present case represents the youngest child with progeria and documented cerebral infarction whose cerebrovascular abnormalities were imaged noninvasively with MRA. The images indicate diffuse cerebral ischemia with superimposed cortical infarcts. The subdural collections were thought to represent old hematomas due to the fragility of bridging veins. Complications of intraarterial angiography have been reported, ranging from 1.3%-12.2% for transient and 0.1%-5.2% for permanent deficits. The higher rates of complication are restricted to individuals with cerebrovascular disease, especially those with anterior circulation transient ischemic attacks and severe stenoses (6, 7). Despite the age differential between our patient and those in published studies, the pathophysiology and his symptoms placed him in the high-risk group. This led to parental preference for MRA over intraarterial angiogra-



A

B

Fig. 3. Intracranial 3-D time-of-flight MRA.

A, Collapsed 0° maximum intensity pixel shows poor intracranial signal, lack of branching vessels of the middle cerebral arteries and absent A1 segment of the right anterior cerebral artery. Ophthalmic arteries are prominent (*arrows*), suggesting collateral circulation.

B, Lateral view confirms the identity of the enlarged ophthalmic artery collaterals (*arrow*). (3-D fast imaging with steady precession 40/13/15°).

phy. The parents consented to repeat MRA because of its noninvasive nature and the use of sedatives rather than general anesthesia.

The various methods of MRA, advantages, and limitations are well documented (8). This case exemplifies the utility of MRA to demonstrate the needed information regarding cerebrovascular disease without risk of an invasive procedure. In summary, we report the third known case of a child with progeria surviving cerebrovascular infarction as a result of premature atherosclerosis. This is the first known use of MRA in this population to demonstrate noninvasively severe atherosclerotic disease.

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